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CITATION:

Sano, Katsuyuki ...[et al]. A case of infant with bilateral multicystic kidney and ureteropelvic junction obstruction; surgical treatment after percutaneous nephrostomy. 泌尿器科紀要 1994, 40(6): 515-518

ISSUE DATE:

1994-06

URL:

<http://hdl.handle.net/2433/115287>

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A CASE OF INFANT WITH BILATERAL MULTICYSTIC KIDNEY AND URETEROPELVIC JUNCTION OBSTRUCTION; SURGICAL TREATMENT AFTER PERCUTANEOUS NEPHROSTOMY

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We report a case of bilateral multicystic kidney and left ureteropelvic junction (UPJ) obstruction in a female infant. She was diagnosed to have severe cystic disease from 20 weeks of gestation. After birth with cesarean section, she was transferred to the neonatal intensive care unit (NICU) of our hospital. One day after birth, she was referred to our department for progression of azotemia. We placed a nephrostomy catheter into the largest renal cyst, but it did not work. One week later, we placed another nephrostomy catheter into another cystic lesion inside of the first one. It worked well for nine months and azotemia was improved. At the age of nine months her upper urinary tract was reconstructed by pyeloplasty. We observed peristalsis of ureter and first urination from bladder.

(Acta Urol. Jpn. 40: 515-518, 1994)

Key words: Multicystic kidney, UPJ obstruction, Nephrostomy

INTRODUCTION

Recently fetal ultrasonography is utilized at many hospitals and diseases with anatomical change of fetus are frequently diagnosed¹⁾. This case was also diagnosed as cystic disease of kidney at the 20th week of gestation. The parents and obstetrician discussed the problem and decided to continue the pregnancy. To prevent fatal dysfunctions, we considered diverse treatment with the agreement of the parents.

Problems that could be fatal were 1) infantile respiratory deficiency syndrome (IRDS), and 2) anuria. The NICU doctors dealt with the first problem by respirator management and pulmonary surfactant. Against anuria, we first tried to drain the large cystic lesions and after several months found that one of the cystic lesions was exactly a dilated renal pelvis. Therefore we tried to repair the normal ureteropelvic alignment by pyeloplasty. Fortunately, the ureter reached the pelvis and we could anastomose the pelvis and ureter.

CASE HISTORY

The patient was a 10-month-old female.

She was the first child of healthy parents, and her mother had a history of natural abortion. When she was in the 20th week of gestation, bilateral cystic lesions were found by fetal ultrasonography. The obstetrician informed her parents on the probability of disease but they decided to continue the pregnancy. At 35 weeks, for preperiodical rupture of gestational bladder and placenta previa, she was delivered by cesarean section on August 11, 1992. The volume of amniotic fluid was very small and the fetus was incarcerated in the pelvis. There was no evidence of anomaly on the body surface. After delivery, she suffered from respiratory deficiency and was transferred directly to our hospital. On admission, blood urea nitrogen (BUN) was 8 mg/dl, and serum creatinine (Cr) was 0.7 mg/dl. There was no evidence of urination. The next day (Aug. 12), BUN, Cr were elevated to 17.6 and 1.64 respectively. We diagnosed her with a multicystic kidney from the computed tomography (Fig. 1) and placed a 3Fr plastic tube under echo guide into the largest cystic lesion as temporary percutaneous nephrostomy. However, urinary output was 100ml for the first day and less than 10 ml there-



Fig. 1. Computed tomography (CT) of abdomen at the day of birth. Over half of the abdomen was occupied with cystic lesions of bilateral kidneys. Left kidney had no perenchyma.



Fig. 2. CT of abdomen a week after the birth, just before second nephrostomy was performed. The largest cyst in Fig. 1 had been drained by first nephrostomy (arrow).

after. BUN, and Cr continued to be elevated up to 46.8 and 5.33 by Aug. 20. We tried to place the second nephrostomy into the deep cystic lesion next to the first one (Fig. 2). The second nephrostomy worked well for a long term and BUN, and Cr were 40.0 and 1.76 by Sept. 3, and 33.5 and 1.03 by Dec. 3. She was attacked by severe respiratory infections several times in those days but after a few months, her respiratory system gained normal resistance against infection. Her neurological and physical development was normal for age. Therefore, we planned the surgical treatment for better quality of life. Retrograde cystography (Fig. 3) revealed the ureter at the level of renal pelvis and the pelvis showed minimal dilatation. As the image of the right kidney was not detectable by renal scintigraphy (^{99m}Tc DMSA), we gave

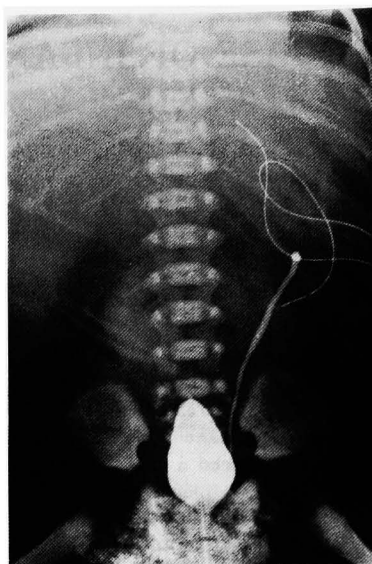


Fig. 3. Retrograde cystography revealed left vesicoureteral reflux and small capacity of bladder.

up its treatment. On the contrary, radioactive uptake with cold defect was detected on the left side.

Cystoscopic findings revealed that the left ureteral orifice was observed in the normal portion with a stadium shape; the inter-ureteric ridge was not clear on the right side, and the right ureteral orifice was not identified.

SURGICAL TREATMENT

From our operative evaluations there were three problems; 1) discontinuity between the left renal pelvis and ureter, 2) the nature of renal pathology, and 3) existence of large cyst at the lower pole of kidney. We planned pyeloplasty, unroofing of the cyst, and renal biopsy. The left kidney was explored through the flank incision, and there was a large cyst on the lower pole. The contents was serous fluid. We performed unroofing and made the biopsy of the edge of the cyst. Then we found the ureter and chased it to the pelvis. The ureter ran on the anterior surface of the pelvis and was in contact to it with fibrous tissue. We resected the excess of pelvis and upper ureter. A 5Fr tube could pass through the residual ureter.

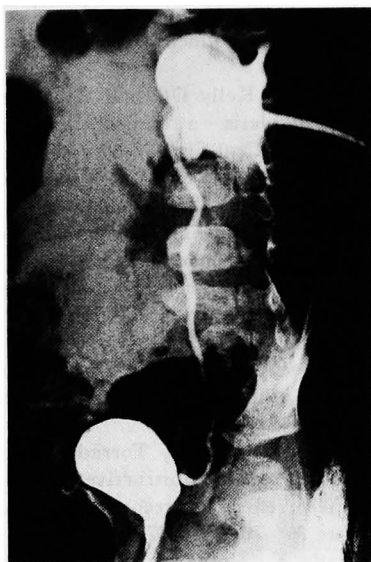


Fig. 4. Nephrostography revealed good passage between kidney and ureter, and peristalsis of the ureter.

Therefore we performed pyeloplasty after the Anderson-Hynes' method. A 5Fr splint tube was left in the ureteropelvic junction and a 14Fr Malecot catheter was indwelt in the pelvis. The splint tube was removed at the 7th day after operation. On the 10th day, we performed nephrostography and confirmed the passage of contrast dye from the pelvis into the ureter, and normal urination. As the patient had a single functioning kidney, we plan to leave the nephrostomy catheter for a long period. After 1.5 months, we performed second nephrostography at catheter change and confirmed the good passage (Fig. 4). However, as intrapelvic pressure was still 35 cm H₂O, we clamped the catheter for 1 hour. With more than 2 hours of clamping, she often developed nausea and bad humor resembling symptoms of urolithiasis.

With the specimen of our renal biopsy, pathologists reported that there was no evidence of renal dysplasia. As we did not obtain any specimen of the right kidney, a pathogenetic difference may exist between the two kidneys.

DISCUSSION

The points of issue are; 1) is the pathogenesis the same albeit there was a large difference in the image diagnosis between the two kidneys? 2) was there any passage between the pelvis and ureter at first, or was there no connection between them? 3) do the ureter and the bladder work well for a long period? and 4) should we select any better treatment from admission to now?

The biopsy specimens from the left kidney revealed atrophic glomeruli and tubuli not typical in renal dysplasia. Quinn et al.²⁾ reported that the primitive duct surrounded by concentric layers of the cellular mesenchyme was an essential criterion of renal dysplasia. We did not find the histological feature in our specimen. Quinn also reported that cartilage may be seen in dysplasia and other diseases, which we could not see.

The cystic lesion that we had punctured first might have been hydrocalicosis (Kellalis et al.³⁾, Shinno et al.⁴⁾), but there was no evidence. Passage between the pelvis and the lesion was not seen at the second nephrostography (Fig. 4). Urine was not secreted from the lesion after catheter removal. Scarce parenchyma remained in the area.

The passage between renal pelvis and ureter was unlikely to exist at birth from radiologic findings. However, without any passage, she would not survive in the uterus. Blane et al.⁵⁾ reported that severe pulmonary hypoplasia, which might be fatal, would associate with bilateral renal obstructive dysplasia. We estimate the period of obstruction to be between 12 and 20 weeks of gestation. As normal organ development begins at about 12 weeks and there were cystic lesions in her kidney at 20 weeks.

As we observed peristalsis of ureter at postoperative nephrostography, the ureter will transport urine effectively to the bladder. Nephrostography 1.5 months after the operation showed better function of ureter, but there remained still high intrapelvic

pressure. This experience suggested that many cases of children's hydronephrosis could improve with age. In this case, we observed improvement of intrapelvic pressure (10 cmH₂O 6 months after the operation), which might be the effect of growth.

Now, we believe that it would have been better to puncture the cystic lesion during her fetal period. Renal development and function would be restored effectively if the obstruction is bypassed before the 20th gestational week^{6,7)}. We recommend early drainage at neonatal and even fetal period as much as possible. As mentioned above, there are many cases of hydronephrosis that improve, with age especially with nephrostomy. However, this case showed no evidence of passage after the treatment, and an operation was indicated.

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(Received on October 25, 1993)

(Accepted on January 24, 1994)

和文抄録

両側の嚢胞腎に腎盂尿管移行部の閉鎖を伴った新生児の1例 経皮的腎瘻を造設した後の手術療法

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両側の嚢胞腎に左腎盂尿管移行部の閉鎖を伴った女児の1例を報告した。この児は20週齢から重篤な嚢胞性疾患を診断されていた。帝王切開によって出生の後、当院のNICUに転送された。高窒素血症のため1日齢で当科に併診された。最大の嚢胞に腎瘻を造設

したがこれが効果なく、1週後にその内側に再度腎瘻を造設した。これが9カ月間有効で高窒素血症は改善した。9カ月齢で腎盂形成術をおこなった。尿管には蠕動が見られ、膀胱から初めて排尿があった。

(泌尿器要 40: 515-518, 1994)